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DOI:

[10.1016/j.prrv.2017.03.002](https://doi.org/10.1016/j.prrv.2017.03.002)

Document Version

Peer reviewed version

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Citation for published version (APA):

Linkson, L., Macedo, P., Perrin, F. M. R., & Elston, C. M. (2017). Anorexia nervosa in Cystic Fibrosis. *Paediatric Respiratory Reviews*. <https://doi.org/10.1016/j.prrv.2017.03.002>

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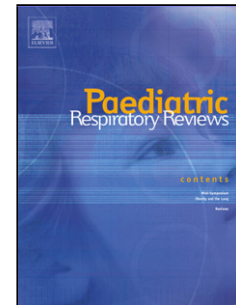
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Accepted Manuscript

Title: Anorexia nervosa in Cystic Fibrosis

Authors: Lynette Linkson, P. Macedo, F.M.R. Perrin, C.M. Elston



PII: S1526-0542(17)30025-8

DOI: <http://dx.doi.org/doi:10.1016/j.prrv.2017.03.002>

Reference: YPRRV 1200

To appear in: *YPRRV*

Please cite this article as: Linkson Lynette, Macedo P, Perrin FMR, Elston C.M. Anorexia nervosa in Cystic Fibrosis. *Paediatric Respiratory Reviews* <http://dx.doi.org/10.1016/j.prrv.2017.03.002>

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Anorexia nervosa in Cystic Fibrosis

Lynette Linkson, P Macedo, FMR Perrin, CM Elston .

Adult Cystic Fibrosis Service, King's College Hospital NHS Foundation Trust, Denmark Hill, London, SE5 9RS

e-mail: lynette.linkson@nhs.net

Summary

This article explores the challenges associated with diagnosing and managing eating disorders such as anorexia nervosa amongst adolescents and adults with cystic fibrosis. It reviews the known risk factors, generic versus disease specific eating disorder risk screening tools and considers the ethical dilemmas associated with critically low body mass indices. A case review is included to illustrate the complexities of managing both conditions in the context of declining respiratory function.

Keywords: Cystic fibrosis; eating disorders; eating behaviours

Introduction

The association between good nutritional status and improved health outcomes in cystic fibrosis (CF) is firmly established (1). It is achieved by regular dietetic review to encourage high calorie diet, adequate pancreatic enzyme replacement, alongside early intervention and investigation of weight loss. The co-existing diagnosis of an eating disorder in CF has the potential to hasten respiratory decline as a consequence of malnutrition.

Case review

This 20 year old lady was diagnosed with CF age 2years 11months on a background of recurrent respiratory tract infections. She is homozygous for the Phe508del mutation, has osteopenia, CF related diabetes (CFRD), and chronic *Pseudomonas aeruginosa* and *Achromobacter xylosoxidans* colonisation. She also has a history of bilateral sensorineural hearing loss secondary to meningitis at age 6 weeks and took a paracetamol overdose age 14 years. On transition to the adult service her forced expiratory volume in one second (FEV1) was between 40 to 50% with a Body Mass Index (BMI) of 21, which had started to drift downwards from the age of 15 years. She has a heavy treatment burden and frequent admissions to hospital with worsening lung function requiring long term oxygen therapy and nocturnal non invasive ventilation (figure 1). In the previous year she had 153 days of intravenous antibiotics. She found it very difficult to accept her declining health. The only thing which she felt able to control was her weight. She was diagnosed with anorexia nervosa age 19 years with a BMI of 17.8 kg/m². Her disordered eating behaviours include diet restriction, variable compliance with pancreatic enzyme supplementation and daily body checking. Her body image is distorted as she believes there is no problem with her weight.

Her BMI fell to 14.8 in April 2015 (figure 2). At that point she had been deemed to lack capacity so could have been detained under the Mental Health Act but agreed to voluntary transfer to the Eating Disorders Unit. She stayed for eight months in the Eating Disorders Unit, with intermittent readmissions to hospital for treatment of pulmonary exacerbations. On discharge from the unit her BMI was 15.2 and her FEV1 remained below 30%. She had gained under one kilogram during that period. She has ongoing input from the community eating disorders team. She had declined enteral feeding until six months ago when she attempted an overnight feed but only tolerated the nasogastric tube for 2.5 hours.

Her glycaemic control has been poor which may be representative of disease progression and frequent infections. It is also a result of insulin omission which may be a direct attempt to manipulate her weight. The CF team has systematically tried to optimise all of her care. For example using tally charts and I-neb downloads to work on her adherence and at her best this reached 100%. She has had regular home visits and outpatient reviews with the multidisciplinary team, which includes a dedicated CF psychologist and diabetologist. She has recently commenced Ivacaftor/lumacaftor.

She would like to improve her BMI in order to be referred for a lung transplant. The eating disorders and CF teams acknowledge that it can take years to successfully treat anorexia nervosa. Her current BMI precludes her from referral for lung transplant. Without a timely improvement in her BMI she may miss the opportunity to be considered for lung transplantation.

Discussion

Anorexia nervosa has the highest mortality amongst psychiatric disorders at three times greater than all other conditions. It is multifactorial in origin and is associated with obsessive, perfectionist and competitive traits. It can also be triggered as a coping mechanism (2). On average it takes five or six years for complete recovery. A third of patients may never recover although there have been reports of recovery 21 years after diagnosis. Anorexia nervosa affects 1 in 250 females and 1 in 2000 males, with a peak incidence occurring in late adolescence and early twenties. Poorer outcomes are associated with a presentation between 20 and 29 years and lower weights.

How can we identify, manage and monitor CF patients at risk of developing eating disorders?

The structure of CF care means patients receive regular clinical review providing an opportunity to monitor BMI and eating patterns. Eating disorders must be considered as a potential cause for unexplained weight loss. Risk factors for developing eating disorders include factors which may be a direct consequence of severe CF disease such as delayed growth and puberty, low body weight, comorbidities such as CFRD, preoccupation with disease and dietary management, as well as reduced life expectancy. Whereas other risk factors such as social isolation, low self esteem and body image disturbance may be related to external influences or as a result of CF(3).

It has been observed that with a declining nutritional status, the perception of body image becomes an increasing concern for adults with CF (4). There are also non eating-related factors such as clubbing or medicalisation of the person by the insertion of a porta Cath, which can also negatively impact on body image (5).

Generic screening tools for eating disorders can be inadequate in the context of CF. Some of the measures of disturbed eating attitudes and behaviours may simply reflect the impact of the condition or its management, for example a preoccupation with food due to dietary recommendations (6, 7, 8). Disordered eating behaviours can exist without the diagnosis of an eating disorder and early recognition of these patterns helps identify those at greater risk. Generic measures fail to assess for disease specific eating attitudes and behaviours such as those related to poor nutritional status, for example feeling full easily. Therefore, a CF specific screening tool has been developed to identify these behaviours, which include omitting pancreatic enzymes or insulin in order to control weight. (5)

It might appear that there should be a greater incidence of eating disorders amongst the CF population given the continued emphasis on the attainment of optimal BMI; however the evidence

is inconclusive (6). In general, adolescents with chronic illnesses that have a dietary component have been found to be at increased risk for developing eating disorders or disordered eating behaviour compared to their peers (6). However, with respect to CF there is some work which demonstrates adolescents and young adults with CF have eating behaviour and attitudes, body satisfaction and self-esteem, similar to healthy controls. There is a suggestion that females with CF actually have fewer problems (5).

It seems within a subset of CF patients there is evidence of an increase in risk factors and disordered eating behaviours. In CFRD, particularly females, there are increased concerns around eating behaviours as well as dissatisfaction with body appearance and low self-esteem. Unsurprisingly adults requiring enteral tube feeding have issues related to body image, self-esteem and have a poorer quality of life. NICE guidance recommends screening for eating disorders in young people with type 1 diabetes and poor treatment adherence so there may be a role for a similar targeted approach within CFRD (9).

The presence of risk factors for eating disorders and disordered eating behaviours identifies those who warrant psychological assessment or closer monitoring. Some of these indicators may be caused by the psychological consequences of treatment burden, disease progression and medicalisation of the body. It may also be that disordered eating behaviours may actually represent attentive and self-regulated eating which helps maintain optimal BMIs (10).

What are the ethical dilemmas of managing critically low BMIs in the context of CF and anorexia nervosa?

Poor nutritional status and low BMI have a significant impact on physical health and in the setting of severe lung disease can be ultimately life threatening. This is a result of respiratory muscle weakness and increased energy requirements. In the context of anorexia nervosa, discussing the risks associated with low weight and respiratory failure does not lead to behaviour change. It is likely that the individual will disregard the significance of such risk, given that ambivalence is an inherent feature of anorexia nervosa. The dilemma in the context of anorexia nervosa and severe respiratory disease is that a BMI below 14 kg/m² has a far more significant impact on physical health than in those without additional comorbidity. Mental Health Act Assessments, to determine capacity, are typically considered when BMIs fall below 14 but in the context of declining physical health this would need to take place sooner.

Without capacity, an individual could be detainable under the Mental Health Act, in order to initiate inpatient treatment for an eating disorder. This is usually only considered as a last resort as it is so restrictive and likely to impact on future therapeutic relationships. Nasogastric tube feeding is not standard practice in eating disorder care and it would only be considered if someone was refusing all oral intake or rapidly declining in weight. Logistically this is difficult to enforce when an individual is physically resistant and feeding against the will of a patient requires legal consideration. This contrasts with the nutritional recommendations in CF where enteral feeding is considered if despite supplementation BMI remains below 20 kg/m² or there is ongoing weight loss (11, 12).

Reviewing the literature (9), there are mixed outcomes for compulsory admission. Potential benefits include: weight gain, reduction in morbidity and mortality, improvement in mood, concentration and symptoms, and reduced cognitive impairment. The potential risks include: barrier to formation of a therapeutic relationship, negative physical and psychological impact, negative countertransference, stigma associated with the Mental Health Act, loss of self-esteem and escalation of resistance.

Cognitive function is impaired by low weight and anorexia nervosa. Usually this improves with weight restoration but in some cases it may not fully recover. This is an important consideration in the management of patients both during and after recovery from an eating disorder and also in CF patients malnourished from other causes.

How do you support patients and the multidisciplinary team to set realistic goals and expectations?

It is essential to maintain a consistent approach across the CF multidisciplinary team, psychiatric community teams and eating disorder units. This might involve regular communication through professional meetings to discuss the best interests of the patient. In order to facilitate admission to an eating disorders unit, the CF team can provide support and training in specialised respiratory care such as physiotherapy, non invasive ventilation and oxygen. During hospital admissions the eating disorder team may visit or liaise with the CF unit to provide ongoing support. The first target for eating disorders is to achieve weight stability. The primary objective of the eating disorders team is to provide a psychological intervention and the duration of treatment and follow up may last years. Weight gain can occur sometime after this has been completed. This is challenging for the CF team as they strive to provide holistic care. The presence of an active eating disorder precludes weight gain and low BMI is a contraindication for lung transplant referral. Therefore, the clinical approach needs to adapt to incorporate the limitations an eating disorder has on disease management, in particular with regard to enteral feeding.

Discordance between patient goals and health related behaviours is a feature of nonadherence and can be seen within eating disorders. It is reported that 50% of medications prescribed for chronic illness are not taken as directed and this can vary with individuals over time (13). It is important to understand this, utilise objective measures and provide ongoing support to enable patients to maintain or improve adherence.

Conclusion

In CF there are complex interactions between health status and psychosocial influences and it remains unclear the extent to which these may be associated with the development of anorexia nervosa (7). It is evident that the impact of weight loss in this condition is far more significant than in the healthy population. It is therefore important to consider eating disorders in patients with CF who have unexplained weight loss to ensure early identification and psychological assessment, particularly given the prolonged recovery time for this condition.

No conflicts of interest.

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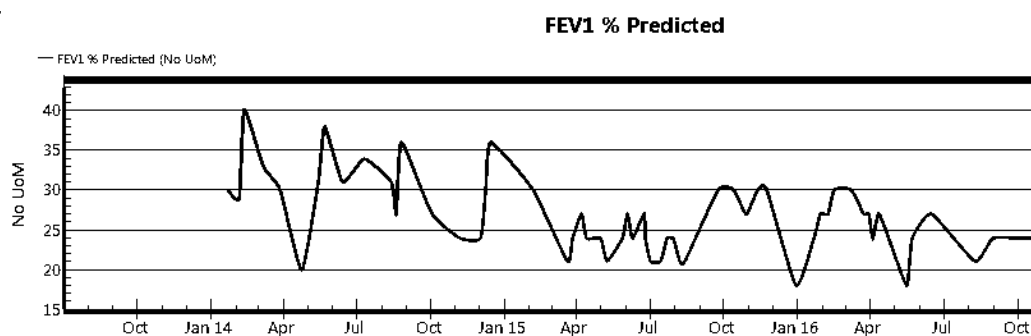


Figure 1

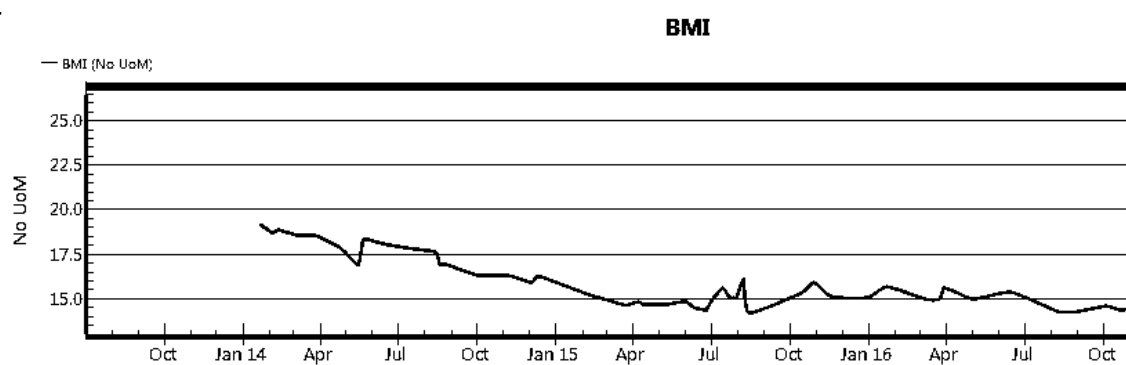


Figure 2